tacks of acute intermittent porphyria (AIP), due to sustained vasospasm, appears to exist.[1] Hyponatremia is a common complication in this disease, that is usually related to a syndrome of inappropriate secretion of antidiuretic hormone (SIADH).[2,3] Rhabdomyolysis may be a frequent complication during hyponatremia correction in which elevation of creatine phosphokinase (CPK) levels runs parallel to the recovery of sodium levels and renal function is not usually impaired.

We report the case of a 16-year-old female patient born in Mauritania, with a family history of acute intermittent porphyria, who was admitted to hospital for abdominal pain and vomiting for the past several days associated to an impaired consciousness level and two episodes of generalized tonic-clonic seizures, after which she remained in a postictal state and required orotracheal intubation and mechanical ventilation. Laboratory test results include severe hyponatremia (Na 95 mM/L; normal, 135-145) associated to plasma (244 mOsm/L) and urinary hyposmolarity (222 mOsm/L), urinary frequency and data suggesting hypovolemia (central venous pressure of 4 mmHg). Isotonic saline infusion was therefore started. Rhabdomyolysis was detected 24 hours after admission. Maximum CPK levels of 35628 U/L were found at 48 hours, but no renal function impairment was noted. Patient clinical signs and history suggested an AIP attack, that was confirmed by measuring in a spot urine sample levels of delta-aminolevulinic acid of 39.7 mg/g (normal, 0-5), as well as values in 24-hour urine of 154.3 mg of porphobilinogen (normal, 0-2), 484 µg/24 h of coproporphyrin (normal, 0-60), and 1,471 µg/24 h of uroporphyrin (normal, 0-22). Once diagnosis was confirmed, treatment was started with intravenous human hemin at 3 mg/kg/day for 4 days. The patient developed arterial hypertension that required administration of beta-blockers. Clinical course was satisfactory, with a progressive improvement in consciousness level. Mechanical ventilation was withdrawn 6 days after admission, and patient was discharged with a normal consciousness level and no symptoms. A genetic study to search for mutations and screen for heterozygous family carriers was requested.

AIP is the most common and severe of hepatic porphyrias.[4] Disease transmission is autosomal dominant, and the gene encoding AIP is located in the long arm of chromosome 11. AIP causes deficiency of the enzyme uroporphyrinogen I synthetase, formerly called porphobilinogen synthetase (PBG), thereby blocking heme synthesis. The most typical clinical signs include nausea, vomiting, constipation, diarrhea, urinary retention, tachycardia, hypertension, mental symptoms, and muscle pain and weakness. These attacks may be triggered by barbiturates, anticonvulsants, estrogens, oral contraceptives, alcohol, or low-calorie diets. Seizures may occur in 20% of cases, particularly in patients with hyponatremia.

The relationship between hyponatremia and rhabdomyolysis may apparently result from intracellular potassium efflux to compensate for the cell edema caused by decreased sodium levels, which would cause a decrease in transmembrane potential and hence in muscle metabolism.[5] Other authors postulate that it is hyponatremia correction itself which causes changes in muscle cells ion concentrations and osmolarity resulting in incapacity to maintain homeostasis in cell volume regulation. This would cause membrane fragility and muscle enzyme release into the bloodstream.[6]

1. Anderson C, Wikberg A, Stegmayr B, Lithner F. Renal symptomatology in patients with acute intermittent porphyria. A population-based study. *J Intern Med* 2000; 248 (4): 319-325.
2. Salgueira M, Monroy A, Jarava C, Molas R, Páez MC, Amor J et al. Hiponatremia en la porfiria aguda intermitente. Un problema diagnóstico en urgencias. *Nefrología* 1997; 17 (1): 88-91.
3. López Montes A, Lorenzo I, Pérez Martínez J. Síndrome de secreción inadecuada de hormona antidiurética y porfiria. *Nefrología* 2004; 24 (Extr 3): 85-88.
4. Anderson KE, Bloomer JR, Bonkovsky HL, Kushner JP, Pierach CA, Pimstone NR, Desnick RJ. Recommendations for the diagnosis and treatment of the acute porphyrias. *Ann Intern Med* 2005; 142 (6): 439-50.
5. Trimarchi H, González J, Olivero J. Hyponatremia-associated rhabdomyolysis. *Nephron* 1999; 82 (3): 274-7.
6. Rizzieri DA. Rhabdomyolysis after correction of hyponatremia due to psychogenic polydipsia. *Mayo Clin Proc* 1995; 70 (5): 473-6.

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Biliptysis as the initial symptom of a rare complication of autosomal dominant polycystic kidney disease

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To the editor: Autosomal dominant polycystic kidney disease (PKD) is an inherited disease usually occurring in adults and characterized by gradual development of multiple renal and extrarenal cysts during life, with different and unpredictable growth rates.[1] This disease occurs in approximately 1 out of every 1,000 people, and is the most common polycystic kidney disease.

This letter discusses the case of a 62-year-old female patient, a Caucasian with chronic renal failure (CRF) secondary to PKD who attended the office complaining of fever for the past 4 days, followed by right chest pain irradiating to hypochondrium and right flank, associated to cough and breathlessness. Some days later she suffered increased breathlessness and cough, which produced an intense yellow expectoration, was sudden, and left an unpleasant taste (biliptysis). The patient experienced right pleural effusion. Minimum undifferentiated pleurotomy was performed, and broad spectrum antimicrobial treatment was administered. She subsequently showed an image with the appearance of a walled abscess in the right base, in addition to pneumothorax, which prompted the decision to perform conventional surgery, at which a biliobronchial fistula from the lower pulmonary lobe and a hepatic cyst, joined together and fibrosed with the phrenic nerve in the fistulous tract area, were found (fig. 1).

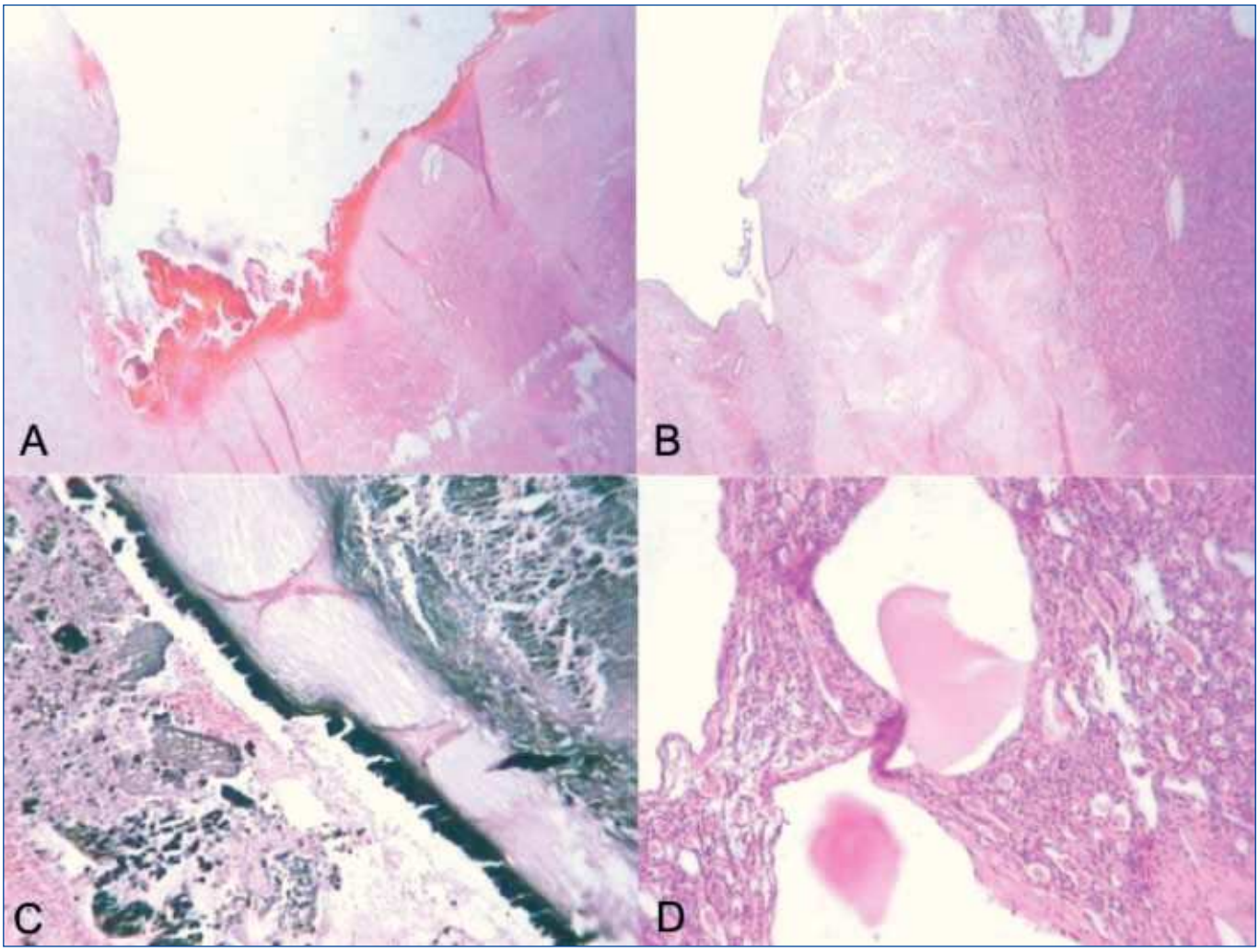

Figure 1. A) Hematoxylin and eosin (HE), 40x, pulmonary tissue. Fistulous tract with presence of bile. B) HE, 40x, hepatic tissue; hepatic cyst, areas of fibrosis and reactive hepatitis. C) 40x, pulmonary tissue, Fouchet method: bile prigments in the fistulous tract. D) HE, cyst in renal parenchyma.

This is the first case reported in Cuba of a patient with PKD having a biliobronchial fistula from a liver cyst that communicated first with the pulmonary parenchyma and subsequently entered the pleural space. The main symptom was biliptysis. A single case has been reported in the literature reviewed.[2] This occurred in a patient with PKD and liver cysts in whom diagnosis was made using high-resolution computed tomography with no contrast agent. Hepatic cysts most commonly reported as causing biliobronchial fistula are those from an infectious origin, such as hydatid and amebic cysts.[3,4] Data on treatment of biliobronchial fistula in the setting of PKD are few or none, because of the rarity of this complication. Drainage through an endoscopically inserted nasobiliary catheter as reported by Partrinou[4], achieving reverse drainage to the biliobronchial fistula by decreasing duodenal pressures as compared to biliary tract pressures, is a noninvasive method that would be the procedure of choice for treating patients at a high surgical risk. The interest of this case lies in that, while this type of complication is uncommon in PKD patients, it should be considered in order to make an accurate diagnosis and start adequate management. It is also recommended that this condition is added to the long list of complications of PKD patients with CRF.

1. Sanz S, Coto E. Enfermedad poliquística: clasificación, etiopatogenia, diagnóstico y actitudes terapéuticas. Cap 12.1. Enfermedades quísticas. En: Avendaño LH. Nefrología Clínica. 2.ª ed. Madrid: Ed. Médica Panamericana; 2003.
2. Bird R, Fagen K, Taysom D, Silverman ED. A case of bronchobiliary fistula in the setting of adult polycystic kidney and liver disease, with a review of the literature. *Clin Nucl Med* 2005; 30 (5): 326-8.
3. Sahin E, Enon S, Cangir AK, Kutlay H, Kavukcu S, Akay H et al. Single-stage transthoracic approach for right lung and liver hydatid disease. *J Thorac Cardiovasc Surg* 2003; 126 (3): 769-73.
4. Partrinou V, Dougenis D, Kritikos N, Polydorou A, Vagianos C. Treatment of postoperative bronchobiliary fistula by nasobiliary drainage. *Surg Endosc* 2001; 15 (7): 758.

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